

## “Ebstein’s Anomaly” a Potential Cause of Misinterpretation on Myocardial Perfusion Imaging

### Abstract

Ebstein’s anomaly is a rare congenital heart disease. We present the case of a 47-year-old woman with history of Ebstein’s anomaly who was referred to nuclear medicine department for a myocardial perfusion single-photon emission computed tomography (SPECT) imaging during pre-anesthetic check-up. Myocardial SPECT perfusion images with (99mTc)-Sestamibi demonstrated medium sizes fixed perfusion defect in the mid and proximal infero-septal wall adjacent inferior wall. This is a cause of potential misinterpretation on myocardial SPECT perfusion imaging as this perfusion defect was due to fibrosis rather than scarred myocardium.

**Keywords:** (99mTc)-Sestamibi, Ebstein’s anomaly, myocardial single-photon emission computed tomography perfusion imaging

A 47-year-old woman with a known history of Ebstein’s anomaly, was referred for myocardial perfusion imaging to our department during her preanesthetic checkup. She was supposed to undergo surgery for carcinoma stomach. The transesophageal echocardiography revealed dilated right ventricle with a 3 cm. Ostium secundum atrial septal defect. There was no pulmonary artery hypertension with a normal pulmonary venous return. The left ventricular function was preserved. A myocardial perfusion single-photon emission computed tomography (SPECT) with a 1-day protocol was performed (stress/rest [99mTc]-Sestamibi). The pharmacological stress test was performed with adenosine infusion for 6 min, with no significant electrocardiogram changes. 10 mCi and 25 mCi of radiotracer was injected during the stress and rest part of the study, respectively. SPECT perfusion images demonstrated a medium-sized fixed perfusion defect in the mid and proximal and mid-inferoseptal wall and adjacent inferior wall [Figure 1]. In addition, the right heart chambers appeared enlarged. To confirm the perfusion defect, represents fibrosis rather than scarred myocardium, coronary angiography. The coronary angiography was found out to be normal. Ebstein’s anomaly is a rare congenital heart disease.<sup>[1]</sup> It involves

the tricuspid valve which is displaced downward toward apical portion of the heart.<sup>[2,3]</sup> This causes atrialization of the right ventricle. There is ventricular hypoplasia and functional impairment. In newborns, it presents as cyanosis,<sup>[4,5]</sup> whereas it may vary from arrhythmia to exertional dyspnea in adults. Ebstein’s anomaly is one of the causes of the misdiagnosis of coronary artery disease. Adult patients with Ebstein’s anomaly develop increased fibrosis in the left ventricular wall and basal and mid-septum.<sup>[6]</sup> This presents as left ventricular regional wall motion abnormalities.<sup>[7]</sup> This is in concordance with the findings of the myocardial perfusion scan in the described case, where the region with decreased counts represents the mid and proximal inferoseptal wall, which is fibrotic and therefore without (99mTc) uptake. The other explanation for the reduced septal perfusion in patients with Ebstein’s anomaly could be the exaggerated motion of the septum or high attenuation caused by an enlarged right atrium leading to decreased counts from the septum.<sup>[8]</sup> Hence, it is imperative for nuclear medicine physicians to be aware of such uncommon but important artifacts while reporting myocardial perfusion scintigraphy.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the

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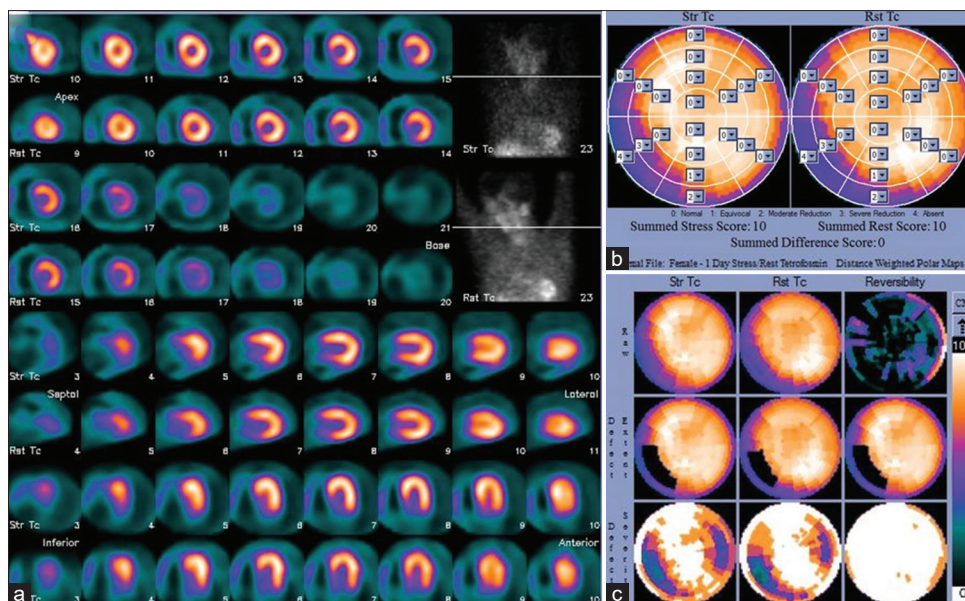


Figure 1: (a) Slices along the short axis, vertical long axis, and horizontal long axis, respectively, (b) summed stress score and summed rest score map, (c) polar maps

form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

#### References

- Attenhofer Jost CH, Connolly HM, Dearani JA, Edwards WD, Danielson GK. Ebstein's anomaly. *Circulation* 2007;115:277-85.
- Schreiber C, Cook A, Ho SY, Augustin N, Anderson RH. Morphologic spectrum of Ebstein's malformation: Revisitation relative to surgical repair. *J Thorac Cardiovasc Surg* 1999;117:148-55.
- Martinez RM, O'Leary PW, Anderson RH. Anatomy and echocardiography of the normal and abnormal tricuspid valve. *Cardiol Young* 2006;16 Suppl 3:4-11.
- Bernstein D. Ebstein anomaly of the tricuspid valve. In: Kliegman RM, Stanton BF, Geme JS, Schor NF, Behrman RE, editors. *Nelson Textbook of Pediatrics*. 20<sup>th</sup> ed., Vol. 2. Philadelphia, PA, USA: Elsevier; 2016. p. 2221-2.
- Cetta F, Dearani JA, O'Leary PW, Driscoll DJ. Tricuspid valve disorders: Atresia, dysplasia, and ebstein anomaly. In: Allen HD, Shaddy RE, Penny DJ, Feltes TF, Cetta F, editors. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult*. 9<sup>th</sup> ed., Vol. 1. Philadelphia, PA, USA: Lippincott Williams & Wilkins; 2016. p. 949-81.
- Frescura C, Angelini A, Daliento L, Thiene G. Morphological aspects of Ebstein's anomaly in adults. *Thorac Cardiovasc Surg* 2000;48:203-8.
- Daliento L, Angelini A, Ho SY, Frescura C, Turrini P, Baratella MC, *et al.* Angiographic and morphologic features of the left ventricle in Ebstein's malformation. *Am J Cardiol* 1997;80:1051-9.
- Grossman BG, Raghuvver KH, Book W, Hunsche A, Santana CA, Garcia EV. Correct interpretation of myocardial perfusion SPECT study in a patient with Ebstein's anomaly through recognition of septal artifact. *J Nucl Cardiol* 2003;10:572-4.